



## Clinical profile, management and outcome of patients diagnosed with Bronchiectasis enrolled in the Chronic Lung Disease Program in a tertiary pediatric hospital from 2021-2023

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**OBJECTIVE:** To describe bronchiectasis profiles at the Philippine Children's Medical Center (2021-2023).

**MATERIALS AND METHODS:** A retrospective chart review at PCMC analyzed the demographic profile, imaging, management and outcome of patients with bronchiectasis enrolled in the Chronic Lung Disease Program from 2021-2023.

**RESULTS:** Twenty seven patients were included in this study. Most patients were diagnosed after the age of six and showed female predominance. The most common symptom at the time of diagnosis was chronic cough (81.5%) with pulmonary tuberculosis (51.9%) as the most common etiology. All patients were treated with cyclic azithromycin. *Pseudomonas aeruginosa* was the prevalent microorganism isolated in the sputum and tracheal aspirate samples of the patients (56.5%).

**CONCLUSION:** Patients with bronchiectasis were more commonly diagnosed in children past age of six with more prevalence in females. The most common symptom was chronic cough followed by fever, dyspnea and weight loss. Pulmonary tuberculosis and recurrent respiratory infections were noted to be the most common etiology with the left lower lobe most affected in HRCT. The most common phenotype seen was cystic, which is irreversible and a sign of progressive bronchiectasis, which may point to a late diagnosis. This emphasizes the need for physicians to have a high index of suspicion in patients with chronic or recurrent respiratory symptoms. All patients were treated with an oral macrolide with 25-31% of patients with decreased and/or absent symptoms. *Pseudomonas aeruginosa* was the prevalent microorganism isolated in the sputum and tracheal aspirate samples of the patients, which should be taken into account when treating for exacerbation.

**KEYWORDS:** *Bronchiectasis, patient profile, treatment outcomes*

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## INTRODUCTION

Bronchiectasis is a syndrome characterized by recurrent or persistent productive cough, airway infection and inflammation, and abnormal bronchial dilatation on chest computed tomography (CT) scans<sup>[1]</sup>, and is mostly the result of an intrinsic airway pathology resulting in the dilatation<sup>[2]</sup>. Studies on pediatric bronchiectasis compared to other respiratory diseases have been scarce, however an increasing prevalence in the pediatric population has been observed in the past years. Globally, it has been estimated that around 0.2 to 735 per 100,000 children are affected annually <sup>[3]</sup>. According to the Philippine Pediatrics Society registry, there are 510 cases recorded as of May 7, 2024 since January 2006, 33 (6.4%) of which came from Philippine Children's Medical Center (PCMC) <sup>[4]</sup>. The Chronic Lung Disease Program in PCMC was established in 2021 for patients with disorders that affect the lungs and respiratory system that usually develop slowly and may get worse over time. Included in the program are patient with bronchopulmonary dysplasia, chronic lung disease of infancy, interstitial lung disease and bronchiectasis who may or may not be on oxygen therapy. The program uses a standardized guide on managing and monitoring patients with these diseases. Due to the lack of research regarding pediatric bronchiectasis, most treatment approaches have been based on adult guidelines. However, studies have found that

mild pediatric bronchiectasis may potentially be reversed or, at the very least, its progression may be stopped with optimal clinical management<sup>[5]</sup>, hence a pediatric treatment approach may be beneficial.

Bronchiectasis is a heterogenous disease that is thought to be a consequence of multiple factors including infectious, congenital, inflammatory and immunologic causes. In a study in 2014 by Brower et al. comprising 989 children with non-cystic fibrosis bronchiectasis, infectious processes such as pneumonia, measles, tuberculosis and pertussis topped the list (17%)<sup>[6]</sup>. The mean age of diagnosis was 8 to 9 years seen across the studies of Lee, et al and Kumar et al. Management of bronchiectasis involves treatment of acute exacerbations, provision of maintenance therapy and evaluation and treatment of underlying causes<sup>[7,8]</sup>. Oral macrolides, particularly azithromycin, showed significant reduction in the exacerbation of non cystic fibrosis bronchiectasis pediatric patients at 24 months<sup>[9]</sup>. Surgical management of bronchiectasis, such as lobectomy, is only considered when maximal medical management has failed and the patient has poor quality of life. As mentioned previously, bronchiectasis when treated in early stages may be treatable and reversible. A study by Gailliard et al. comprising of 32 pediatric patients diagnosed with bronchiectasis with repeat imaging after a median of 24 months showed complete resolution in six patients undergoing medical treatment.

Eight patients showed improvement in repeat imaging. Among the 32 patients, 16 showed resolution of bronchial dilation<sup>[10]</sup>. Generally, the prognosis greatly varies depending on the severity of the disease on the time of diagnosis, host's response to treatment and etiological factors. Delayed diagnosis has been associated with poorer outcomes. Other poor prognostic factors include comorbidity of asthma, bilateral lung involvement, saccular type of bronchiectasis, increased frequency of exacerbations and presence of *P. aeuruginosa* in the airways<sup>[11,12]</sup>. In line with this, early diagnosis with optimal management and prevention of complications would increase the chances of reversibility and improve prognosis, especially in the pediatric population.

This study aims to present the clinical profile, management and outcome of patients with bronchiectasis at Philippine Children's Medical Center from the year 2021 to 2023. The age, sex, etiology identified, and symptoms at the time of diagnosis were included. The management and monitoring parameters done during the 1st, 2nd and 3rd year follow up including macrolide use, surgery, sputum microbiology, pulmonary function tests and 2D echocardiography were obtained, as well as the number of exacerbations, rehospitalizations and mortality.

## **MATERIALS AND METHODS**

The study was conducted as a case review of patients with the diagnosis of

bronchiectasis obtained from the Chronic Lung Disease Program (CLDP). The study included patients less than 18 years old with the diagnosis of bronchiectasis seen at the CLDP of PCMC from 2021-2023. Patients who were not diagnosed via high resolution Chest CT scan (HRCT) were excluded from the study. All patients included in the list of patients with the diagnosis of bronchiectasis obtained from the CLDP were included.

Data was collected from the record of the CLDP, as well as from the inpatient and outpatient charts from the Medical Records. The clinical profile, including the age, sex, etiology, symptoms and microbiologic profile, of each patient was recorded. Diagnostic modalities including high resolution chest CT scan was recorded. Treatment done during their admissions as well as outpatient was also included. Monitoring using lung function test, symptom monitoring, exacerbations, rehospitalizations and mortality was recorded. Data was presented as mean (SD) or number (%) as appropriate.

## **RESULTS**

A total of 27 children aged less than 18 years old diagnosed with bronchiectasis through HRCT and enrolled in the CLDP from 2021-2023 were included in the study. Four patients (14.8%) were diagnosed with bronchiectasis prior the establishment of CLDP, hence were enrolled once CLDP was established in 2021.

Two (7.4%), twelve (44.4%) and nine (33.3%) of the patients were diagnosed in 2021, 2022 and 2023, respectively.

Table 1 shows the demographic parameters of these patients. Most of the patients were diagnosed past the age of six with a male to female ratio of 1:1.7. More than half of the patients (59.2%) had comorbidities such as cerebral palsy, gastroesophageal reflux disease and bronchial asthma. Most patients presented with chronic cough (81.5%) followed by fever (66.7%), dyspnea (63%) and weight loss (59.3%) (Table 2).

Table 1. Demographic Profile of patients diagnosed with Bronchiectasis enrolled in the Chronic Lung Disease Program

	Demographic Parameters	Cases n (%)
Age at time of diagnosis	0 to <2 years	1 (3.7)
	2-5 years	4 (14.8)
	6-10 years	10 (37.0)
	>10 years	12 (44.4)
Gender	Male	10 (37.0)
	Female	17 (63.0)
Co-morbidities	Cerebral Palsy	4 (14.8)
	Gastroesophageal reflux disease	3 (11.1)
	Bronchial Asthma	3 (11.1)
	Congenital heart disease	2 (7.4)
	Central Hypoventilation Syndrome	2 (7.4)
	Focal Epilepsy	2 (7.4)
	Connective tissue disease	1 (3.7)
	Systemic Lupus Erythematosus	1 (3.7)
	Interstitial Lung Disease	1 (3.7)
	Congenital Hypothyroidism	1 (3.7)
	Diabetes Mellitus	1 (3.7)
	Tracheomalacia	1 (3.7)

Most cases had an etiology identified for the cause of bronchiectasis, of which the most common was pulmonary tuberculosis (51.9%). Other etiologies noted were recurrent pneumonia (25.9%), endobronchial tuberculosis (11.1%) and interstitial lung disease (3.7%) (Table 3).

Table 2. Symptom at the time of diagnosis

Symptom at the time of diagnosis	Cases n (%)
Chronic cough	22 (81.5)
Fever	18 (66.7)
Dyspnea	17 (63.0)
Weight loss	16 (59.3)
Recurrent Respiratory Infection	9 (33.3)
Easy fatigability	6 (22.2)
Hemoptysis	6 (22.2)
Increased sputum production	4 (14.8)
Body malaise	2 (7.4)
Chest pain	1 (3.7)
Clubbing	1 (3.7)

The distribution of bronchiectasis as seen in the HRCT of the patients enrolled in CLDP showed majority of patients with pulmonary tuberculosis to have a unilateral lower lobe distribution (42.9%). There was an almost equal distribution in patients with pulmonary tuberculosis that presented with cylindrical (57.1%) and cystic (42.9%) changes. While in recurrent pneumonia, there was nearly equal distribution between unilateral and bilateral involvement, still with predominantly lower lobe affected.

Three patients each among the seven patients (42.9%) with recurrent pneumonia had cylindrical and varicose type of bronchiectasis. All patients with endobronchial tuberculosis had left lower lobe cystic bronchiectasis (Table 4 and 5).

Figure 1 shows the microbiologic flora obtained from the sputum and/or tracheal aspirate of the patients associated with each diagnosis. The most common organisms cultured are *Pseudomonas aeruginosa* (56.5%), and *Klebsiella pneumoniae* (34.8%).

Table 3. Etiology of Bronchiectasis

Etiology	Cases n (%)	Age at the Time of Diagnosis
Pulmonary Tuberculosis	14 (51.9)	9.5 (2-17)
Recurrent Pneumonia	7 (25.9)	6 (0.9-16)
Endobronchial Tuberculosis	3 (11.1)	11 (6-16)
Idiopathic	2 (7.4)	13 (8-18)
Interstitial Lung Disease	1 (3.7)	8

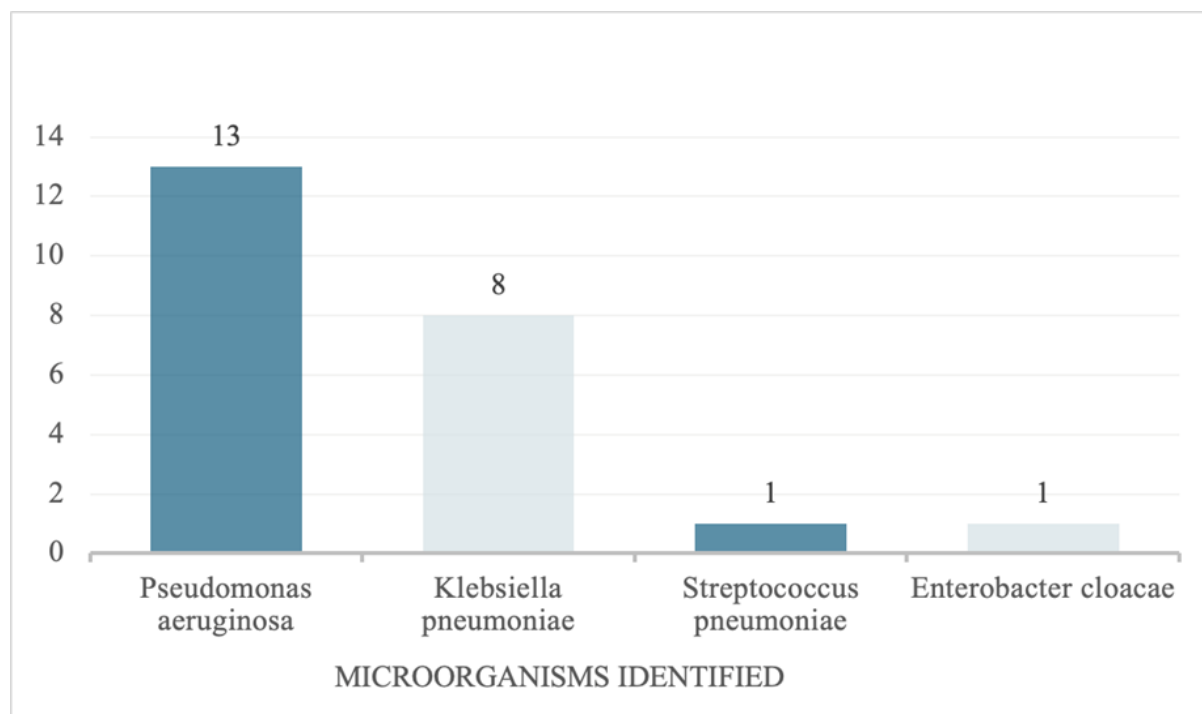
Table 4. Lobar Involvement of Bronchiectasis

Etiology	UL	ML	LL	R	L	B	W
Pulmonary Tuberculosis	2	2	6	2	7	2	3
Recurrent Pneumonia	3		6	3	1	3	
Endobronchial Tuberculosis			3		3		
Idiopathic			2		2		
Interstitial Lung Disease			1			1	

UL, upper lobe involvement; ML middle lobe involvement; LL lower lobe involvement; R, right lung; L, left lung; B, bilateral; W, widespread – involvement of 4 or more lobes

*Table 5. Phenotype of Bronchiectasis*

<b>Etiology</b>	<b>Cylindrical</b>	<b>Varicose</b>	<b>Cystic</b>
Pulmonary Tuberculosis	8		6
Recurrent Pneumonia	3	3	1
Endobronchial Tuberculosis			3
Idiopathic			2
Interstitial Lung Disease			1



*Figure 1. Microorganism identified in sputum/tracheal aspirate of bronchiectasis patients*

All patients diagnosed with bronchiectasis were started on cyclic azithromycin at 30mg/kg/week. Seven patients were referred for thoracocardiовascular surgery (TCVS) however conservative management was recommended. This may be attributed to the improvement of symptoms on cyclic azithromycin, presence of comorbidities or decision of the family. One patient referred to TCVS also transitioned to adult care. 2D echocardiography was done in 18 patients (66.7%), two of which (7.4%) had elevated PAP. Both had severe pulmonary hypertension at PAP by TR jet 98mmHg and 81mmHg.

They were referred to Cardiology service and was subsequently managed with sildenafil for the pulmonary hypertension.

Patients included in the study were enrolled to the CLD program from 2021 to 2023. Eighteen patients enrolled to the CLD program prior to 2023 were eligible for the 1<sup>st</sup> year follow up while only six patients enrolled prior to 2022 were eligible for the 2<sup>nd</sup> year follow up and only four patients enrolled prior to 2021 were eligible for the 3<sup>rd</sup> year follow up. Nine patients were enrolled in 2023 hence follow up was not included in the study.

*Table 6. Outcome on the 1st year follow up*

		Cases n (%)
<b>Improved symptoms</b>	Decreased cough frequency	5 (31.2)
	No cough	2 (12.5)
	Weight gain	2 (12.5)
<b>Exacerbation treated as outpatient n=7</b>	Increased sputum production	5 (71.4)
	Increased cough frequency	4 (57.1)
	Fever	4 (57.1)
	Dyspnea	1 (14.3)
<b>Exacerbation needing rehospitalization n=6</b>	Increased sputum production	5 (83.3)
	Increased cough frequency	4 (66.7)
	Fever	4 (66.7)
	Dyspnea	6 (100.0)
	Seizure breakthrough	1 (16.7)

*Table 7. Outcome on the 2nd year follow up*

		Cases n (%)
<b>Improved symptoms</b>	Decreased cough frequency	1 (25)
	No cough	0
	Weight gain	0
<b>Exacerbation treated as outpatient n=2</b>	Increased sputum production	2 (100)
	Increased cough frequency	2 (100)
	Fever	1 (50)
	Dyspnea	2 (100)
<b>Exacerbation needing rehospitalization n=3</b>	Increased sputum production	2 (66.7)
	Increased cough frequency	3 (100)
	Fever	3 (100)
	Dyspnea	3 (100)
	Hemoptysis	1 (33.3)



Table 6. Outcome on the 3rd year follow up

		Cases n (%)
<b>Improved symptoms</b>	No cough	1 (33.3)
<b>Exacerbation treated as outpatient n=1</b>	Increased sputum production	1 (100)
	Increased cough frequency	0
	Fever	0
	Dyspnea	0
<b>Exacerbation needing rehospitalization n=2</b>	Increased sputum production	2 (100)
	Increased cough frequency	1 (50)
	Fever	0 (0)
	Dyspnea	2 (100)

Sixteen of eighteen patients (88.9%) came in for follow up at 1 year of diagnosis at the outpatient department (Table 6). One patient (5.6%) transitioned to adult care after passing the age of 18. Five patients (31.3%) reported decrease in cough frequency while two patients (12.5%) claimed to have no cough at all. Two patients (12.5%) also reported weight gain from previous consults. However, seven patients (43.8%) had at least one exacerbation treated with antibiotics at the OPD while six patients (37.5%) had at least one exacerbation needing rehospitalization. In patients with exacerbation treated at the OPD, the most common symptom was increased sputum production (71.4%) followed by increased cough frequency (57.1%). Co-amoxiclav was given in five patients (71.4%) while Ciprofloxacin was given to two patients (28.5%) with symptom relief. The same symptoms was seen in patients needing rehospitalization due to bronchiectasis exacerbation, however of note is that all

patients had dyspnea. One person was treated with vancomycin, gentamicin, ceftriaxone and clindamycin while the another patient was given vancomycin and ciprofloxacin. The remaining four patients were given piperacillin-tazobactam. There was no reported mortality.

Six patients diagnosed prior to 2022 were eligible for the 2<sup>nd</sup> year follow up (Table 7). Four of six patients (66.7%) were seen for their 2<sup>nd</sup> year follow up, three of which were hospitalized for acute exacerbation (75%). Two of the patients were given piperacillin tazobactam while all patients were given ciprofloxacin. One patient had recurrent hospital acquired infection hence was also given meropenem, levofloxacin, co-amoxiclav, gentamicin and clindamycin. Two patients (50%) also had exacerbations treated as outpatient and were given co-amoxiclav. One patient was transitioned to adult care (16.7%). There was no reported mortality.



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Four patients (diagnosed prior 2021) were eligible for the 3<sup>rd</sup> year follow up (Table 8). Three of four patients (50%) were seen for their 3<sup>rd</sup> year follow up. One (33.3%) had improved symptoms however two (66.7%) had exacerbations needing rehospitalization one given ceftriaxone and the other piperacillin tazobactam. In total, out of the 27 patients included in the study, there was no reported mortality during the 3-year period.

## DISCUSSION

Bronchiectasis is a chronic respiratory syndrome characterized by recurrent or persistent productive cough, airway infection and inflammation, and abnormal bronchial dilatation on chest computed tomography (CT) scans<sup>[1]</sup>. Bronchiectasis in the pediatric population, particularly those not caused by cystic fibrosis, has previously been thought of as a rare disease, however recent studies have shown increasing prevalence worldwide. Around 0.2 to 735 per 100,000 children are affected annually[3] with prevalence and

severity increasing with age. The Chronic Lung Disease Program (CLDP) in the Philippine Children's Medical Center (PCMC) listed 27 patients diagnosed with bronchiectasis since its establishment in 2021 until 2023. The mean age of diagnosis is 9 years, which was congruent with the studies of Lee, et al. and Kumar et al, which comprised of 387 and 80 patients, respectively. However, there is a female predominance of 1.6:1 in our study compared to the male predominance of both studies.

Bronchiectasis has three main phenotypes – tubular, varicose and cystic. Among these, tubular bronchiectasis has been most identified with high resolution computed tomography (HRCT)<sup>[13]</sup>. However, cystic bronchiectasis was the predominant phenotype identified in patients with bronchiectasis enrolled in CLDP. This is significant as studies show cylindrical bronchiectasis is considered to be the mildest form of bronchiectasis. As the disease progresses, irregularities form in the bronchial wall with reduction of identifiable bronchi as the bronchi becomes varicose in characteristic with focal constrictive areas between the dilated airways. The most severe form is considered the cystic phenotype, which was most the major phenotype seen in our study, wherein there is progression of the dilatation forming grape like clusters. The least number of bronchi is seen in cystic types, now with involvement of the terminal bronchioles<sup>[11]</sup>. A case report on

an adult patient with bronchiectasis following acute infection with initial findings of cylindrical changes showed negative results on repeat CT scan after 10 months. However, reversal was only seen in cylindrical and saccular types and not in cystic types<sup>[15]</sup>. The results may point to a late diagnosis, which may be explained by social determinants such as poor health seeking behaviors, financial constraints and inadequate resources. A study by Kawi, et al on Filipino's health seeking behaviors. showed only 69.1% primarily sought consult with a doctor with others turning to herbal medications, prayers and visiting rural health clinics<sup>[25]</sup>. This may be seen as a barrier to early diagnosis which may explain why majority of the patients enrolled in CLDP presented with a progressive form of bronchiectasis. However, the CLDP database lacks information on the sociodemographic profile of patients' families, which could be valuable in identifying potential barriers to effective medical management. Family-related sociodemographic factors play a significant role in healthcare outcomes, as they can impact a patient's access to care, understanding of medical instructions, and overall treatment adherence. These factors include the 1.) Family's socioeconomic status which can influence their ability to afford medications, treatment, or follow-up appointments, potentially creating barriers to optimal care; 2.) Cultural and Language Differences which may affect how well they understand medical advice, follow treatment guidelines, or

communicate with healthcare providers, which can hinder care delivery; 3.) Educational Level which can impact their ability to grasp complex medical information, follow care instructions, and manage treatment regimens; 4.) Social Support or the availability of family or community support which can significantly affect the patient's ability to adhere to treatment, attend appointments, and manage day-to-day care needs and 5.) Access to Healthcare Resources which can determine their access to necessary healthcare services, such as transportation, specialists, and follow-up care. Without this crucial sociodemographic data, healthcare providers and researchers may miss important insights that could improve personalized care, identify challenges early, and ultimately enhance patient outcomes<sup>[26,27]</sup>.

Bronchiectasis is a heterogeneous disease that is thought to be a consequence of multiple factors<sup>[16]</sup>. A myriad of etiologies have been described in bronchiectasis including infectious, congenital, inflammatory and immunologic causes. Post infectious causes were determined to be the etiology in our study with pulmonary tuberculosis the most common etiology of bronchiectasis. In a study by Lee et al. comprising of 387 patients in secondary and tertiary hospitals in South Korea, majority (55.3%) also showed post infectious causes as the etiology of bronchiectasis. However, pulmonary tuberculosis only comprised of 12.3%, which may be attributed to the socioeconomic

differences. A different study done also in South Korea showed an identified etiology in 86% of cases with bronchiolitis obliterans (33%), mostly post infectious, as the leading cause, followed by childhood respiratory infection including tuberculosis (21%)<sup>[17]</sup>. Interstitial lung disease, immunodeficiency, primary ciliary dyskinesia and cystic fibrosis were also identified. Bronchiectasis can be divided into noncystic fibrosis bronchiectasis (NCFB) and cystic fibrosis bronchiectasis (CFB). Although majority of cases are not caused by cystic fibrosis, noncystic fibrosis bronchiectasis has only been relatively studied in the recent years compared to that of cystic fibrosis bronchiectasis. Due to the rare occurrence of the gene affected in cystic fibrosis, particularly transmembrane conductance regulator (CFTR) gene located on chromosome 7 in the Asian population, prevalence is low and in turn, bronchiectasis attributed to cystic fibrosis is less than that of Western countries. In the Philippines, there have been no studies on the prevalence of the disease in the country, however a case report by Almonte, et al. presented the first case of a Filipino infant to be diagnosed with cystic fibrosis<sup>[18]</sup>. Bronchiectasis in the Philippines therefore is mostly attributed to non-cystic fibrosis causes.

In patients with bronchiectasis, increased sputum production leads to recurrent or persistent productive cough, a key symptom of bronchiectasis. Patients also experience dyspnea, wheezes, fever and hemoptysis<sup>[17]</sup>.

Chronic productive cough unresponsive to at least 4 weeks of oral antibiotics should prompt investigation as they are at significantly greater risk of having radiographic changes of bronchiectasis<sup>[19]</sup>. A study done by Wurzel et al showed 1 in 12 patients with protracted bacterial bronchitis developing bronchiectasis at 2 years with recurrent episodes of protracted bacterial bronchitis and lower airway infection with *Haemophilus influenzae* as the major risk factors<sup>[20]</sup>. This is compatible with the findings in our study wherein chronic cough is present in 81.5% of the patients diagnosed with bronchiectasis. Fever, dyspnea, weight loss and hemoptysis were also seen, which is consistent with the most common etiology being pulmonary tuberculosis.

The goals of management in patients with bronchiectasis comprises of preventing further airway damage and premature respiratory decline, optimizing the patient's quality of life, minimizing exacerbations and preventing complications<sup>[21]</sup>. As discussed earlier, in some children, early identification and treatment may halt progression and even render bronchiectasis reversible. Maintenance therapy includes airway clearance techniques, mucoactive agents in selected patients with high daily symptoms, frequent exacerbations, difficulty in expectoration and decreased quality of life, and use of chronic antibiotics. Long term use of antibiotics is recommended in reducing the risk of exacerbations in those who had more than one hospitalized exacerbation or three or more non-hospitalized

exacerbations in the previous 12 months. Oral macrolides, particularly azithromycin, showed significant reduction in the exacerbation of non-cystic fibrosis bronchiectasis pediatric patients at 24 months<sup>[9]</sup>. Surgical management of bronchiectasis, such as lobectomy, is only considered when maximal medical management has failed and the patient has poor quality of life. It is generally indicated in patients with severe localized disease and/or recurrent hemoptysis that can be addressed by resection of a bronchiectatic lobe<sup>[21,22]</sup>. In our study, cyclic azithromycin was given to all patients while surgical management was considered in 25.9% of the patients. On follow up during the 1<sup>st</sup> and 2<sup>nd</sup> year, 25% and 31% of patients showed decrease in symptoms with absence of exacerbations, respectively. However, there is a noted increase in exacerbations during the 2<sup>nd</sup> year follow at 2022, which may be attributed to the loosening of the pandemic restrictions nationwide, where in people go out more with no masks and there is increase in health seeking behaviors.

Monitoring disease progression in patients with bronchiectasis helps in the early detection of acute exacerbations hence promoting timely intervention, noting the rate of disease progression for prognostic purposes, and measuring the response to given therapies. This may be done through symptom tracking with diary cards, measurement of the sputum volume, lung imaging through CT scan, measurement of lung function through spirometry, tracking of microbiologic profile

of the patients through sputum cultures, and inflammatory biomarkers such as sputum neutrophil elastase. Part of the monitoring plan for patients enrolled in CLDP are symptom monitoring, lung function tests every 3-6 months, sputum testing every 6-12 months and chest CT scan when indicated, however due to financial constraints and unavailability of imaging at the institution, most were not facilitated. Different factors affecting the health seeking behaviors of the guardians of the patients also comes into play why these were not followed through. Sputum and tracheal aspirate cultures showed prevalence of *Pseudomonas aeruginosa* and *Klebsiella pneumoniae* in our study. This is contrast with the study of Lee et al. wherein the most prevalent microorganism cultured was *Haemophilus influenzae*. However, a study by Qin et al. showed similar results with *Pseudomonas aeruginosa* being the most commonly isolated microorganism in both baseline and follow up studies in 30 patients [23].

Pulmonary arterial hypertension is one of the complications in chronic lung diseases including bronchiectasis, hence 2D echocardiography is usually offered 3-6 months. At the time of diagnosis, two patients in our study had elevated pulmonary arterial pressure and had to be maintained on sildenafil. Both also had readmissions due to symptoms of right sided heart failure. In a study by Ocal et al analyzing the effect of

pulmonary hypertension with the long term survival in patients with bronchiectasis, patients with pulmonary hypertension was noted to be more hypoxemic with greater number of involved lobes in HRCT. They associated the finding of pulmonary hypertension with worse survival in bronchiectatic patients<sup>[24]</sup>.

## CONCLUSION

Bronchiectasis in children is most commonly diagnosed after age six, with a higher prevalence in females. The most frequent symptoms include chronic cough, fever, dyspnea, and weight loss. Pulmonary tuberculosis and recurrent respiratory infections are the leading causes, with the left lower lobe most often affected on HRCT. The cystic phenotype, indicating irreversible and progressive bronchiectasis, is commonly seen, suggesting the possibility of a late diagnosis. This highlights the importance of a high index of suspicion in patients with chronic or recurrent respiratory symptoms. All patients were treated with oral macrolides, resulting in a 25-31% improvement in symptoms. *Pseudomonas aeruginosa* was the predominant pathogen isolated from sputum and tracheal aspirate, which should be considered in the management of exacerbations.

## LIMITATIONS OF THE STUDY AND RECOMMENDATIONS

The study covered 27 patients enrolled in CLDP, including their demographics, management and monitoring. However, as seen in other studies, social determinants may affect the time to diagnosis and monitoring of the illness. We recommend including these as they may become barriers in early diagnosis, implementation of effective interventions and providing the optimal management for the patients. The study is also a retrospective chart review and there may be incomplete or missing documentation. This also only focuses on the admissions and consults in our institution, and there may be patients who may also have been seen and/or admitted at other institutions.

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